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Frontotemporal Dementia: Identification and Management

Leah Wilfong, MS, AGPCNP-BC, Nancy E. Edwards, PhD, ANP-BC, Karen S. Yehle, PhD, FAHA, and Karla Ross, MSN, ANP

ABSTRACT

Dementia is a disease primarily affecting older adults in their ability to carry out daily activities. Frontotemporal dementia (FTD), a leading cause of early-onset dementia, can present in the fourth and fifth decade by progressive changes in personality, affect, and behavior. The atypical presentation of FTD may resemble a midlife crisis, depression, schizophrenia, or a traumatic brain injury among its causes. The etiology of FTD is unknown, although genetic links have been suggested, thus treatment focuses on behavioral and symptom management. Early diagnosis of FTD is needed to better support the emotional and financial burdens of family and caregivers.

Keywords: advanced practice nurse, dementia diagnosis, frontotemporal dementia, management

INTRODUCTION

Dementia is a neurodegenerative disease presenting as a moderate to severe decline in cognition with the potential to hinder daily functioning.[1] The leading cause of dementia is Alzheimer's disease (AD), followed by dementia with Lewy body and frontotemporal dementia (FTD).[2] FTD represents about 10% of all diagnosed dementia cases, and is a leading cause of early-onset dementia.[3] The purpose of this article is to improve early diagnosis and management of FTD by increasing awareness, providing education to support primary care providers.

FTD is often mistaken for AD (Table 1), and has been diagnosed as schizophrenia or bipolar affective disorder due to its presentation early in life.[4] AD presents primarily as short-term memory or language loss displayed as forgetfulness, problems finding the right words, difficulty identifying familiar objects, and inability to complete routine tasks.[5] FTD presents with changes in affect, personality, and social conduct, often sparing memory loss in early stages. This is often noted as apathy, antisocial behavior, loss of inhibition, and lack of insight.[2]

Early diagnosis of FTD by clinicians is imperative. Life expectancy from the initial onset of symptoms averages 6.6-9 years, but only 3-4 years from time of diagnosis.[6] The average age of diagnosis is 57 years old, roughly 13 years earlier than the average for AD.[3]

EPIDEMIOLOGY

In the United States, FTD affects an estimated 20,000-30,000 people, with prevalence rates ranging from 15 to 22 per 100,000 in individuals between the ages of 45 and 65.[6] Sixty percent of FTD cases are diagnosed in individuals between the ages of 45 and 64, with an additional 10% presenting at $<$ 45 years of age and 30% presenting at $>$ 65 years of age.[6,7]

RISK FACTORS

The most prominent risk factor is a positive family history of FTD. Family history is found in 30%-50% of individuals[8] and, of these, 40% display an autosomal dominant inheritance pattern.[9] Three genes have been associated with autosomal dominant FTD. Over 80% of hereditary associated FTD is linked to mutations in these genes.[10]

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Table 1. Differentiating Clinical Presentation of FTD from AD[5,19]

	FTD	AD
Age of onset (years)	45-65	65 and older
Progression	Insidious onset and gradual progression	Insidious onset and gradual progression
Area of brain involved	Frontal and temporal lobes	Hippocampus, posterior temporal and parietal regions, cerebral and cerebellar cortex
Cognitive deficits	None; disinterest presents at decreased cognition in later stages	Diminished executive function, impaired visuospatial function, inability to recognize or identify objects despite intact sensory function
Memory	Relatively intact recall about present and past	Initially, remote memory intact, difficulty with new learning and remembering day-to-day events
Behavior	Prominent personality changes, socially inappropriate and poor judgment, apathy, impulsiveness, personal neglect, excessive eating	Paranoid and delusional thoughts, hallucinations, agitation, anxiety, depression, mistaken beliefs
Language	Word-finding and object-naming difficulty, echolalia, gradual reduction in speech culminating in mutism	Word-finding related to memory, echolalia, inability to follow conversation, may progress to muteness
Motor	Repetitive motor behaviors, mild akinesia and rigidity	Mild akinesia and rigidity, impaired ability to carry out motor activities despite intact motor function

AD = Alzheimer's disease; FTD = frontotemporal dementia.

DIFFERENTIAL DIAGNOSIS

Diagnosis of FTD may initially be suspected with reports of dramatic personality, emotional, or behavioral changes. These symptoms can mistakenly be attributed to a midlife crisis, marital conflict, stress, or even menopause. Obtaining a thorough personal and family medical and psychological history is needed. Family members may not have been formally diagnosed with FTD, so inquiring about unusual behavior, unexpected divorces, spending sprees, or legal trouble may provide an indication of FTD presence.

If FTD is suspected, other medical and psychiatric conditions must first be ruled out. Possible medical differential diagnoses include: cerebrovascular accident; delirium; infection; hypothyroidism; vitamin B_{12} and folate deficiencies; head trauma; or brain tumor. Psychiatric conditions, such as bipolar disorder, depression, personality disorder, schizophrenia, and substance abuse, must also be eliminated.[4] Other types of dementias must also be excluded.

DIAGNOSTIC CRITERIA

Diagnostic criteria for FTD were first established in 1994. The most recent set of criteria was published in 2011 by the International Behavioral Variant FTD Criteria Consortium, and outlined in Table 2.[4] These criteria, developed by experts in the field, are the

Table 2. Diagnostic Criteria for bvFTD and PPA[4,6,14]

	PPA	
bvFTD (must meet 3 or more criteria)	Inclusion Criteria (all must be present)	Exclusion Criteria (all must be absent)
1. Behavioral disinhibition 2. Apathy or inertia 3. Loss of sympathy or empathy 4. Compulsive, ritualistic, or stereotyped behavior 5. Hyperorality and dietary changes 6. Executive deficits with intact memory and visuospatial functions	1. Language impairment is most prominent symptom 2. Aphasia is the primary deficit in early stages 3. Language impairment is causative factor for impairments in daily living	1. Early and profound visuoperceptual, episodic, and visual memory deficits 2. Early, prominent behavior disturbance 3. Psychiatric diagnosis accounts for cognitive impairment 4. Medical or neurocognitive disorder accounts for deficits

bvFTD = behavior variant frontotemporal dementia; PPA = primary progressive aphasia.